Case 16095
Double Aortic Arch

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Section: Cardiovascular
Published: 2018, Sep. 25
Patient: 95 year(s), male

Clinical History

A 95-year-old man, admitted to our hospital for erysipelas, developed progressive drowsiness. Relevant medical history of the patient included arterial hypertension, COPD and type 2 diabetes. No apparent causes for the drowsiness could be identified in the history and clinical examination of the patient.

Imaging Findings

Plain radiography of the chest (Fig. 1) showed an enlarged upper mediastinum with a broad soft tissue shadow at the right side of the trachea (arrowheads). Also remark a left aortic calcification (arrow).

CT angiography of the neck vessels (Fig. 2a and 2b) showed diffuse atherosclerosis, without significant stenosis. A structural anomaly of the aortic arch with two blood vessels encircling the trachea and the esophagus was visualized.

Using 3D-reconstruction techniques of the CT angiography (Fig. 3), a double aortic arch (DAA), right-dominant with in-between the trachea and the esophagus, was visualized. Both on the right and left aortic arch arise the subclavian artery and the carotid artery, with subsequent absence of the right brachiocephalic trunk.

Discussion

BACKGROUND

The incidence of a vascular ring anomaly of the aortic arch accounts for 1-2% of all congenital diseases of the heart and aorta [1]. A double aortic arch (DAA) may be right dominant (70%), as is seen in this case, left dominant (25%) or balanced type (5%) [2]. The formation of a vascular ring, characterized by a connection between the left and right aortic arches, enclosing the trachea and esophagus, may be explained by the failure of the normal regression of one or more segments of the six pairs of the aortic arches [3].

CLINICAL PERSPECTIVE

The resultant vascular ring can cause varying degrees of airway and esophageal compression. A case series of 81 patients showed that the majority of patients with DAA experience symptoms, most often respiratory (stridor) and gastrointestinal (dysphagia) as a result of the indentation of trachea and esophagus [4]. As this rare condition is most often diagnosed at young age, our case is remarkable because of the old age of the patient, and the absence of any clinical symptoms.

IMAGING PERSPECTIVE

Radiography of the thorax shows a right and left paratracheal shadow, representing the right and left aortic arch. Absence of an air column in the thoracic part of the trachea is not present in this case. Other imaging techniques include barium swallow studies, which can be indicated in symptomatic patients to visualize an impression on the esophagus as well as echocardiography to identify associated cardiac anomalies, which are present in a minority of cases. These studies were not performed in this patient, as no clinical signs of compression were present. Although CT angiography was performed to detect a vascular cause of drowsiness, it was also the best choice for the visualization and characterization of the vascular ring. When DAA is suspected in a symptomatic patient, MRI may be the modality of choice because of its unique ability to differentiate vessels from avascular masses, without the need for contrast material or radiation exposure, especially in younger patients.

OUTCOME

In this case, the diagnosis of DAA was made by coincidence in late adulthood, with the patient being asymptomatic during his entire lifetime. However, most patients develop respiratory/gastrointestinal symptoms at young age. In those cases, surgical repair is warranted. Outcomes are usually excellent after repair, although persistent respiratory symptoms are possible, depending on maldevelopment of the trachea and/or major airways, due to previous compression.

Written informed patient consent for publication has been obtained.

Final Diagnosis

A double aortic arch (DAA), right-dominant

Differential Diagnosis List

Other types of aortic arch anomalies, Right arch/left ligament, Aberrant right subclavian artery,

Figures

Figure 1 Figure 1: Plain Radiography (AP) of the thorax.

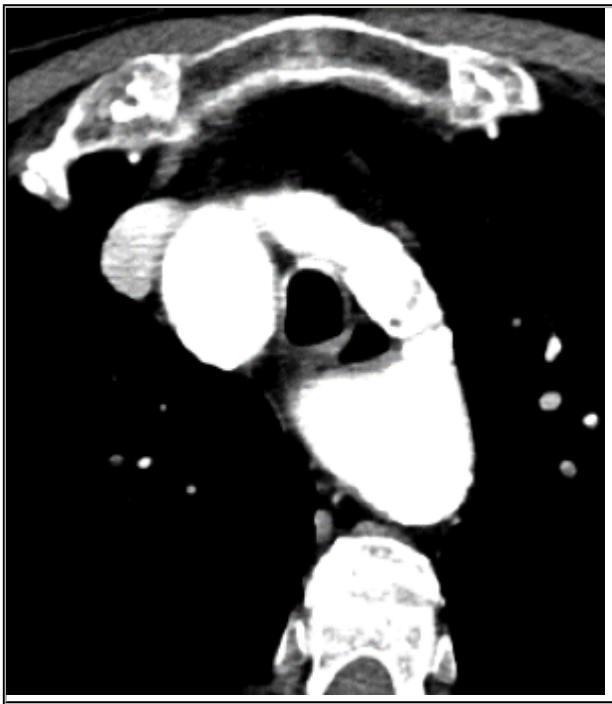
Remark the enlarged upper mediastinum with a broad soft tissue shadow at the right side of the trachea (arrowheads). Also remark a left aortic calcification (arrow).

Area of Interest: Thorax;
Imaging Technique: Plain radiographic studies;
Procedure: Diagnostic procedure;
Special Focus: Arteriovenous malformations;

Figure 2 CT angiography, axial images through the upper mediastinum.

There is a structural anomaly of the aortic arch with two blood vessels encircling the trachea and the esophagus

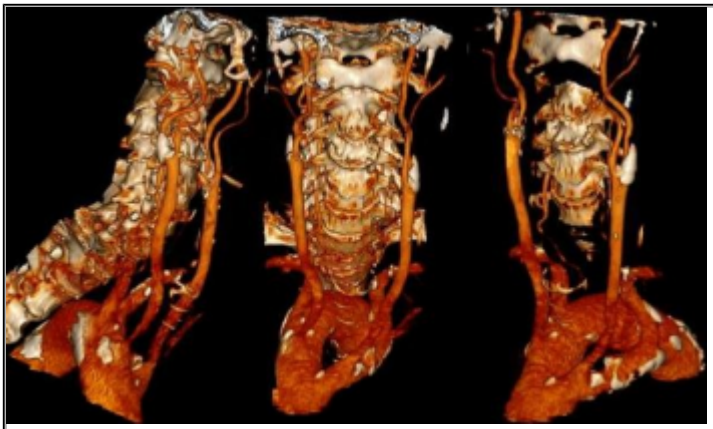
Area of Interest: Arteries / Aorta; Cardiovascular system; Vascular;
Imaging Technique: CT; CT-Angiography;
Procedure: Contrast agent-intravenous; Diagnostic procedure;
Special Focus: Arteriovenous malformations;



There is a structural anomaly of the aortic arch with two blood vessels encircling the trachea and the esophagus

Area of Interest: Vascular;
Imaging Technique: CT; CT-Angiography;
Procedure: Contrast agent-intravenous; Diagnostic procedure;
Special Focus: Arteriovenous malformations;

Figure 3 Figure 3: 3D-shaded surface display of the CT angiography.



Using 3D-reconstruction techniques, the double aortic arch and its branches are easily recognized.

Area of Interest: Arteries / Aorta; Cardiovascular system; Vascular;
Imaging Technique: CT; CT-Angiography;
Procedure: Computer Applications-3D; Diagnostic procedure;
Special Focus: Arteriovenous malformations;

References

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Citation

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Double Aortic Arch {Online}
URL: http://www.eurorad.org/case.php?id=16095